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ANGIOCARDIOGRAPHY IN DISEASE WITH AN ABSENCE OF PULSE

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- USSR -

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ANGIOCARDIOGRAPHY IN DISEASES WITH AN ABSENCE OF PULSE

-USSR-

[Following is the translation of an article entitled "Angiokardiografiya pri bolezni otsutstviya pul'sa" (English version above) by V. V. Zhavoronkov and Ya. V. Paktovskiy (Kuybyshev) of the Clinic of Hospital Therapy (Chief, Prof A. I. Germanov) and the Chair of Roentgenology and Radiology (Chief, Prof Ye. L. Kevesh) of the Kuybyshev Medical Institute in Klinicheskaya Meditsina (Clinical Medicine), Vol XXXVIII, No 5, Moscov, 1960, pages 121-126.]

The ailment of absence of pulse was first described by the Japanese ophtalmologist Takayashi as a casuistic disease accompanied by a lowering of the sharpness of vision. Not so long ago there still existed the wide-spread opinion that this disease is observed only in Japan. However, in 1954 Ask-Upmark, on the basis of literature data and his own observations, gave a summary table of 28 cases of this disease in other countries. At present, the cases of absence of pulse already number about 120, only half of them having been recorded in Japan. In particular, this disease was described in 1955-1958 in the USA, France, Germany, England and Czechoslovakia (R. B. and R. W. Kalmansohn; Friese and Roetzler; Mathieu, Hadot, Pernot and Metz; De Bakey, Morris, Jordan and Coulie; Schwartz and Tomsi; Jedlicka and Urban; Korbelar and Rosol; Horak and Iserle).

From Takayashi's ailment one must distinguish a number of other diseases accompanied sometimes by absence of pulse, such as obliterating endarteriitis, temporal arteriitis, Reno disease, and syphilitic affections of the vessels. Moreover, in differential diagnosis it is necessary to take into account atheromatosis, mediastinal tumors and thromboses, which are also sometimes accompanied by absence of pulse.

The etiology of the ailment of absence of pulse is unknown. Antelava and Byukhem [Buechem?] attach significance to syphilis;

Ask-Upmark and Ch'ang Hsio-t'eng class it among the infectious-allergic diseases or the group of collagenoses, such as nodular periarteriitis, acute red lupus, rheumatism, rheumatoid polyarthritis, serous disease, obliterating thrombangiitis (trombangiit) and temporal arteriitis. The ailment of absence of pulse differs from these diseases in that it affects the large arteries issuing from the aorta, is accompanied by phenomena of ischemia of the upper half of the body and also lacks such symptoms as lingering high fever and a multiplicity of affections of the skin, joints, lymphatic nodes, kidneys, liver, spleen and serous membranes. The clinical symptoms of the disease (Ask-Upmark) depend upon ischemia of the upper part of the trunk, the development of the collaterals, change in the condition of the carotid sinus and cardiovascular reactions.

The disease, as a rule, affects women aged 20 to 30 (rarely older) and is characterized by a progressing course; in some cases brief remissions are possible. Prognosis is unfavorable, and thus far no effective treatment has been proposed. Vessel-expanding drugs, ACTH and cortisone effect some improvement in the condition. Surgery is sometimes applied.

We have observed two patients suffering from Takayashi's disease.

Female patient B, aged 27, was observed in the clinic of hospital therapy of the Kuybyshev Medical Institute from June, 1957, to February, 1959. Her heredity was not burdened. Among diseases suffered, she notes measles (in childhood), tonsillitis and grippe. In 1957 she underwent appendectomy, and in 1959 a tonsillectomy. She considers herself to have been ailing since February, 1957, when after frequently repeated tonsillitis daily chills suddenly appeared, accompanied by a rise in temperature to 38.5°, by rheumatic pain in the joints of arms and legs, severe headaches and abundant secretion of sweat. In July, 1957, these symptoms were joined by vertigo; her temperature rose to 39.5°. In December, 1957, stabbing pains appeared in the region of the heart with irradiation into the left shoulder-blade, palpitation of the heart and shortness of breath when walking. In June, 1958, the patient began to note a chilliness in her upper and lower extremities, a rapid tiring of the arms and a periodic feeling of numbness in them. In July, 1958, the patient began to be alarmed by constant pains along the course of the vasculonervous bundles in arms and legs and along the course of the intercostal arteries, and by vertigo when turning the head and getting out of bed, spots before the eyes and periodic ringing in the ears. In August, 1958, were first noted a slight filling and tension of the pulse in the radial arteries. Arterial pressure was 105/60 mm. During the next ten days the pulse in these arteries could hardly be felt. On 12 August the pulse in the radial arteries of both arms

disappeared, the arterial pressure in the shoulder arteries was not defined, and oscillations were absent. The pulse in the arteries of the back of the foot was distinctly defined, the arterial pressure was 155/80 mm according to the sphygmomanometer, but 250/30 on the oscillogram; the oscillations were pronounced. The pulse in the temporal arteries palpitated. The temporal pressure was 85 mm in the lying position and 100 mm in the standing position (normally, the arterial pressure drops somewhat in this position).

Upon examination, the patient's integuments were found to be pale; the vascular network was well developed in the upper half of the trunk. The upper and lower extremities were cold to the touch. The heart was widened 1 cm. to the left. Over the aorta was heard a systolic noise and the accent of tone II with a metallic timbre. The systolic noise was heard in the vessels of the neck and in the interscapular space. The neck vessels pulsated visibly. The remaining internal organs showed nothing special. Blood analysis: Hb 60 units, er. 4,200,000, color index 0.71, l. 15,6000, e. 2%, p. 3%, s. 67%, lymph. 23%, mon. 5%: ROE 63 mm per hour. Urine without pathological changes. Wasserman reaction and sedimentary reactions negative. Residual nitrogen, cholesterol, bilirubin, blood sugar, non-hemoglobin iron in normal limits. Van den Berg reaction indirect.

In an electrocardiogram made in October, 1958 there was noted a tendency of the electric axis of the heart to incline toward the left, and not very sharply pronounced changes in the myocardium. A small narrowing of the arteries was detected on the part of the bottom of the eye.

Roentgenoscopy and roentgenography of thorax (October, 1958): Lung fields clear, vascular picture of lungs without visible changes. Right cupola of diaphragm somewhat deformed (welding process), sinuses free. Heart without visible changes. In the direct tomograms of the thorax taken at a depth of 9 and 10 cm from the plane of the table (with the patient lying on her back), one distinctly perceives a displacement of the trachea toward the right and a widening of the shadows of both <u>trunci brachiocephalici</u> (Fig. 1). [Note: Illustrations not reproduced here due to poor quality. Captions are appended.] This widening is especially easy to see in the direct tomograms of the neck, taken at a depth of 11 cm from the plane of the table (Fig. 2). In the tomograms taken in left lateral projection at different depths one notes a narrowing of the retrosternal space due to a widening of the ascending aorta.

Diagnosis: ailment of absence of pulse.

Treatment of the patient was made with cortisone (3,250 mg altogether), penicillin (intramuscularly and internally (total amount 30 g), pelentan, pachycarpin, riboflavin, vitamin B_1, ascorbic acid, nicotinic acid and pine-needle baths (37°). In the process of

treatment, the prothrombin activity was maintained at a level of 35%.

On 21 October, 1958 (the 5th day since the beginning of treatment and the 70th day since the disappearance of the pulse, a pulse with slight filling and tension again appeared in the radial arteries of both arms; arterial pressure was not determined; oscillations were poorly pronounced in the right arm and were absent in the left. Arterial pressure in the lower extremities had dropped from 250/80 to 180/80; the temporal pressure had not changed. In the following days the pulse palpitated more distinctly in the right upper extremity than in the left. The general condition of the patient had improved; chills had ceased, the temperature had dropped to normal figures, vertigo had diminished and the patient had taken on weight.

The ROE had slowed to 11 mm an'hour; in the peripheral blood the number of leucocytes had diminished to 9,6000 and the prothrombin activity had dropped to 81.8%. The other indices had not undergone any substantial changes.

From the foregoing history of the disease it may be seen that symptoms typical of the ailment of absence of pulse gradually began to appear on the background of the patient's clinical pictrue, which suggested sepsis. The pathological process apparently affected the subclavian and carotid arteries, resulting in their partial obliteration.

Female patient A, aged 32. Heredity not burdened. At 10 years of age, patient had typhoid, at 11 she contracted malaria. At 18 she had sepsis. From 30 on she had frequent attacks of tonsillitis.

Against a background of complete health the patient at the age of 16 suddenly lost consciousness for a short time. In this same period, shortwindedness occurred when she walked fast. However, the patient continued to perform her usual work and engage in physical culture. At the age of 18 repeated loss of consciousness was noted. In the following years she often fell ill with grippe. At 22 the shortness of breath became intensified, for which reason the patient consulted a physician, who first noted the absence of pulse in both arms. In 1949, after lifting a weight, the patient had a miscarriage; the uterine hemorrhage ceased quickly and independently. At 25 the shortness of breath and weakness became more pronounced, with periodic stabbing pains first appearing in the region of the heart with irradiation into the left shoulder blade. The pulse was absent in the radial arteries. At that time the doctors conjectured the presence of a heart defect in the patient. In 1952 and in 1955 she was treated at a hospital for intensified vertigo, shortness of breath, pains in the rgion of the heart, rapid tiring and weakness. At 27 she lost consciousness for the third time. Three years later she was recognized as a group III invalid. In May and October of 1958 she was examined in the faculty surgical clinic of the Kuybyshev Medical Institute, where a diagnosis of stenosis of the pulmonary artery was

made. She declined the proposed operation.

In November of 1958, at the age of 32 the patient entered the hospital therapeutic clinic for examination with complaints about vertigo when changing the position of her body, the appearance of "circles" before her eyes, periodically occurring ringing of the ears, a feeling of numbness in the arms, a pronounced shortness of breath during walking, rapid tiring, periodic stabbing pains in the region of the heart with irradiation into the left shoulder blade, and palpitation of the heart.

The patient is of regular build and satisfactory nutrition. Integuments and mucous membrane of the lips are cyanotypic. Extremities cold to the touch. Shortness of breath. Heart widened in all directions (principally to the right), tones muted, systolic noise and accent of tone II heard over pulmonary artery, systolic noise over aorta and especially distinct at Bokin's point. Systolic noise also heard in vessels of neck, in subclavian regions and in interscapular space. Pulse does not palpitate in radial arteries of both arms, arterial pressure not determined, oscillations absent. Pulsation in all vessels of lower extremities unsatisfactory. Pressure in artery of back of foot 120/90 mm on right, 150/80 mm on left; oscillations with small pulse-wave amplitude. Temporal pressure 80 mm in lying position; 85 mm in standing position. Blood analysis: Hb 84 units, er. 4,570,000, color index 0.93, l. 6,400, e. 4%, s. 56%, p. 4%, lymph. 31%, mon. 5%; ROE 10 mm an hour. Urine without pathological changes. Wasserman reaction and cytocholic (tsitokholevaya) reaction negative. Residual nitrogen, cholesterol, bilirubin, plasma albumin, non-hemoglobin iron and fibrinogen in normal limits. Van den Berg reaction indirect. Prothrombin index 110%. In the electrocardiogram are noted declination of the electric axis of the heart to the right, pronounced diffusive changes in myocardium with occurrences of chronic coronary failure. On the bottom of the eye a certain undulation and small narrowing of the arteries are noted; veins insignificantly widened, dark in color. Laringologist's conclusion: chronic tonsillitis.

Roentgenoscopy and roentgenography of thorax (May, 1958). Lung fields clear; vascular picture of lungs poorly pronounced. Diaphragm movable, sinuses free. Heart enlarged in dimensions in both ventricles. Aorta without visible changes. Cone of pulmonary artery very bulging. Roentgenokymography of heart: denticles of left ventricle without special changes. Amplitude of denticles of aorta and pulmonary artery somewhat lowered, their height increased. Amplitude of denticles of right contour considerably increased; ventricular component predominates among its denticles.

Probing of heart and angiocardiography: A probe introduced into the elbow vein passed through the subclavian vein, the vena anonyma and the upper vena cava into the right auricle, thence into

The right ventricle, the main introduction of the "cardiotrast" (kardiotrast), the latter is detected in these vessels and in the right auricle. In all the following photos are noted a sharp widening of the cone of the pulmonary artery and a long delay of the contrast substance in it. In a photo taken six seconds after introduction of the "cardiotrast", the contrast substance still fills the cavities of the right heart and the cone of the pulmonary artery. There are indistinct traces of cardiotrast in the vessels of the lungs. Tomography of the heart and of the large vessels (November, 1958), made in direct, left and right lateral projections, confirmed the enlargement of the cavities of the heart, and also revealed widening and deformation of both trunci brachiocephalici (Figs. 3 and 4).

Thus, in the first observation there was affection of the major vessels issuing from the arc of the aorta; in the second observation, the pulmonary artery had also been drawn into this process in addition to these vessels. The congenital character of the affection of the pulmonary artery is gainsaid with a certain amount of probability by the fact that the patient reached the age of 33 without surgery and has continued down to the present time to work in production. This is confirmed also by anamnesis (symptoms of affection of the arterial trunks appeared earlier than symptoms of stenosis of the pulmonary artery). The angiocardiographic data (indistinct traces of contrast in the vessels of the lungs) permit one to think also of affection of the smaller pulmonary vessels, in addition to the main trunk and principal branches of the pulmonary artery, since with isolated affection of the valves of the pulmonary artery the branches of the latter in the lungs are outlined finely but sharply (I. Littman and Z. Fono). At the same time, taking into consideration the data available in the literature, one must suppose that in infectious-allergic process underlies the vascular affections in our patients (both had suffered from a septic condition). From this viewpoint it is possible to admit that the pathologic process causing the symptom complex of Takayashi's disease in this case also affected the vessels of the system of the pulmonary artery, which agrees fully with the pathologo-anatomical data of certain authors (Shimizu and Sano).

Regarding Takayashi's disease as one of the types of collagenosis (Ask-Upmark) and drawing an analogy with nodular periarteriitis of the pulmonary artery, we are inclined to consider the widening of the latter a result of the raising of the resistance of the pulmonary arterioles without any congenital heart defect, just as occurs in nodular periarteriitis (Chi Kong Liu and Lima).

It should be noted that in the roentgenologic examination of our two patients we observed no symptom of the wasting away of the ribs, such as noted by a number of authors in this disease.

At the same time, in a layerwise roentgenologic investigation of the major vessels we succeeded in obtaining a representation of the widened trunci brachiocephalici, which is a direct symptom of Takayashi's disease as it is of panarteriitis.

The improvement in the health of the first female patient after active medicinal (medikamentoznoy) therapy received in the initial stage of the disease, i.e., before distrubance of the brain and eye circulation, testifies to the possibility of temporary restoration of the passability of the affected vessels.

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CAPTIONS TO ILLUSTRATIONS

Fig. 1. Tomogram of the thorax of female patient B, taken at a depth of 9 cm from the plane of the table. Visible in the picture is the displacement of the trachea toward the right and widening of the trunci brachiocephalici.

Fig. 2. The same female patient. Tomogram of the region of the neck, taken at a depth of 11 cm from the plane of the table. Diagram shows bilateral widening of the trunci brachiocephalici.

Fig. 3. Tomogram of the thorax of female patient A, taken at a depth of 9 cm from the plane of the table.

Diagram: 1 - left auricle; 2 - right auricle; 3 - widened and deformed right truncus brachiocephalicus; 4 - bulging cone of pulmonary artery.

Fig. 4. Same female patinet. Tomogram of region of neck, taken at a depth of 10 cm from plane of table.

Diagram: 1 - larynx; 2 - branches of upper-lobe arteries of lungs; 3 - widened and lengthened left great carotid artery.

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